217 Early referral to a specialised centre is associated with a better respiratory outcome

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Background: Specialised CF care is long recognised as a major prognosis factor. Yet, published evidence supporting it remains limited and is often hampered by comparisons with historical controls.
Hypothesis: Could there be differences in outcome variables between children referred to specialised centres less than 2 years after the diagnosis and those referred later?
Methods: In this retrospective multicentric study, data from all CF children fulfilling the following criteria were collected: (1) Age 6 – <18 at the end of 2003; (2) diagnosis before 8 y; (3) follow-up in one of the 6 main CF Belgian centres; (4) at least 1 spirometry and respiratory culture available for 2003. Group A included children referred ⩾2 years after the diagnosis. Each patient from group A was then matched with a single early referred patient on the basis of 3 criteria: same sex, same centre, as closest age as possible (Group B).
Results: Data from 218 children were collected (Group A: 61/218 = 28%, 29M/32F). Results are reported in the table.

	Group A	Group B	p
Age (y), m±SD	13.4±3	13±2.7	NS
ΔF508/ΔF508 (%)	61	61	
Median age at diagnosis (y)	0.2	0.2	
Median delay diagnosis/referral (y)	6.9	0.2	<0.0001
Weight (median Pc)	26	31	NS
Height (median Pc)	36	44	NS
FEV1 (% pr.), m±SD	76±21	86±19	<0.05
% *Pseudomonas aeruginosa* (end 2003)	43	28	<0.05

Conclusion: In Belgium, at age 13, children referred to a specialised CF centre less than 2 years after the diagnosis have a significantly better FEV1 and a lower prevalence of PA than those referred later. Important differences in mean FEV1 were observed between centres (range: 74–95% pred.). This study also provides indirect evidence supporting neonatal screening

218 Long term non-invasive positive pressure ventilation is associated with a stabilisation in the decline of lung function

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Aims: Several short term studies have demonstrated that non invasive positive pressure ventilation (NPPV) is able to improve alveolar hypoventilation and gas exchange during wakefulness, sleep, physiotherapy and exercise in Cystic Fibrosis, but long term benefits have not been demonstrated. The aim of the study was to evaluate the benefit of NPPV in patients with severe lung disease after one year of treatment.
Methods: Patients from the national CF registry who were started on NPPV (n = 53) were compared to matched controls (n = 53), one year before the initiation of NPPV, during the year of NPPV initiation, and after one year of NPPV treatment. Each patient of the NPPV group was matched with a control patient for gender, CFTR genotype, age ± 1 yr, weight ± 2 kg, FEV1 ± 10% predicted, and follow up in the same CF centre. For the 2 groups, the following parameters were compared one year before the start of NPPV (year −1), the year of NPPV initiation (year 0), and one year after the start of NPPV (year +1): weight, body mass index, vital capacity (VC), FEV1, arterial blood gases, number of intravenous (IV) antibiotic courses.
Results: At year −1, the 2 groups were comparable with regard to age, nutritional status, genotype, VC, FEV1, PaO_2, $PaCO_2$, the number of IV antibiotic courses and days with IV antibiotics. At year 0, the NPPV group had a significantly greater decline in VC and FEV1, more IV antibiotic courses. At year +1, the decline in VC and FEV1 was similar in the 2 groups, demonstrating a stabilisation of lung function decline in the NPPV group.
Conclusion: These data show for the first time that NPPV is associated with a stabilisation of the decline in lung function in patients with severe lung disease.

219 Pulmonary function and growth in children with CF in relation to *Pseudomonas aeruginosa* (PA) status and treatment regimen: an observational study

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Aim: To compare pulmonary function and growth in relation to PA antibiotic susceptibility status and treatment regimen.
Methods: Patients attending a single tertiary centre but managed by different antibiotic regimen (Group A, age 0.8–18.2 y: elective 3-monthly IV antibiotics; Group B, age 0.4–17.3 y: rescue antibiotics) were studied retrospectively. They were subdivided by PA status: no PA, PA with no antibiotic resistance (s-PA) and PA with resistance (r-PA) on in-vitro testing using Modified Stokes' method (to ceftazidime, meropenem, tobramycin, piperacillin, colistin and/or ciprofloxacin) from Jan-Mar 2004. Outcome measures were: best FEV_1 (in those >7 y age) expressed as % predicted; height (Ht), weight (Wt) and BMI expressed as standard deviation scores (SDS) using UK 1990 standards.
Results: The distribution of Gp A patients by PA status did not differ from gp B ($P = 0.4$). Gp A patients with no PA had higher Ht and Wt SDS than gp B but FEV_1 and BMI did not differ (Table). Gp B patients with r-PA had lower FEV_1 than gp A (Table).

	No PA		s-PA		r-PA	
	Grp A n = 48	Grp B n = 42	Grp A n = 22	Grp B n = 12	Grp A n = 26	Grp B n = 16
FEV_1 %*	97 (64–146)	90 (48–123)	87 (41–114)	79 (39–111)	80[1] (34–104)	60[1] (33–110)
BMI SDS†	0.1 (1.3)	−0.3 (1.0)	−0.3 (1.1)	0.1 (1.0)	−0.5 (1.1)	−0.3 (1.1)
Wt SDS†	−0.1 (1.2)[2]	−1.0 (1.2)[2]	−0.4 (1.3)	−0.6 (1.1)	−1.1 (1.5)	−1.1 (1.5)
Ht SDS†	−0.3 (1.1)[3]	−1.1 (1.3)[3]	−0.4 (1.3)	−1.0 (1.1)	−1.1 (1.4)	−1.2 (1.4)

*median (range); †mean (SD); [1] $P = 0.04$; [2] $P < 0.001$; [3] $P = 0.002$.

Conclusion: Compared to rescue treatment, an elective treatment regimen was associated with better pulmonary function in patients with antibiotic resistant PA. However, the clinical benefits of an elective regimen did not include better nutritional state and linear growth.

220 Non-invasive ventilation (NIV): experience of Cystic Fibrosis Center of Genova

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Non-invasive ventilation (NIV) includes all techniques that do not need a tracheal prothesis; NIV revolutioned the management of patients (pts) with respiratory insufficiency, reducing use of invasive mechanical ventilation and its related complications. In literature several articles deal about NIV in adults, but not in pediatric pts. NIV has been used in Cystic Fibrosis (CF) pts awaiting for lung transplantation (LT).
We describe our experience in 2 CF pts:
R.C., age 17, was awaiting for LT since Oct 2004. In Nov 2004, after a worsening of the clinical conditions with acute hypoxic hypercapnic respiratory failure, was started to Biphasic Positive Airway Pressure finalized to overcome acute event. NIV, in association with conventional therapies, produced a progressive improvement of clinical conditions (reduction of cough and dyspnoea) and a decrease of venous hypercapnia (101.5 mmHg vs 71.2 mmHg). She continued NIV overnight reporting a remission of awaking symptomatology related to the nocturnal hypercapnia (headache and weakness). In Jan 2006, R. received LT.
F.F., age 15, is waiting for LT since Oct 2005. In Nov 2005, after the begininng of parenteral nutrition, F. felt headache, dispnoea and severe hypercapnia; we decided to start NIV with low pressure (Ipap 14 cm H2O, Epap 2 cm H2O, Pressure Support mode) because of a pneumothorax (PNX) without clinical symptoms, occurred 10 months before. An improvement of the clinical symptomatology was observed immediately. 20 days after discharge, F. was admitted for dyspnoea and severe desaturation. X-ray showed left PNX.
Conclusions: NIV is an important treatment of CF pts awaiting for LT. Past PNX is not an absolute controindication to NIV, but it could be helpful for small time during acute events.